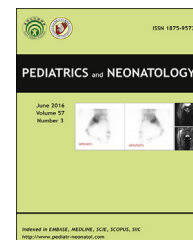


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REVIEW ARTICLE

Longitudinal Follow-up of Chronic Pulmonary Manifestations in Esophageal Atresia: A Clinical Algorithm and Review of the Literature

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tracheoesophageal
fistula

In the past decades improved surgical techniques and better neonatal supportive care have resulted in reduced mortality of patients with esophageal atresia (EA), with or without tracheoesophageal fistula, and in increased prevalence of long-term complications, especially respiratory manifestations. This integrative review describes the techniques currently used in the pediatric clinical practice for assessing EA-related respiratory disease. We also present a novel algorithm for the evaluation and surveillance of lung disease in EA. A total of 2813 articles were identified, of which 1451 duplicates were removed, and 1330 were excluded based on review of titles and abstracts. A total of 32 articles were assessed for eligibility. Six reviews were excluded, and 26 original studies were assessed. Lower respiratory tract infection seems frequent, especially in the first years of life. Chronic asthma, productive cough, and recurrent bronchitis are the most common respiratory complaints. Restrictive lung disease is generally reported to prevail over the obstructive or mixed patterns, and, overall, bronchial hyperresponsiveness can affect up to 78% of patients. At lung imaging, few studies detected bronchiectasis and irregular cross-sectional shape of the trachea, whereas diffuse bronchial thickening, consolidations, and pleural abnormalities were the main chest X-ray findings. Airway endoscopy is seldom included in the available studies, with tracheomalacia and tracheobronchial inflammation being described in a variable proportion of cases. A complete diagnostic approach to long-term respiratory complications after EA is mandatory. In the presence of moderate-to-severe airway disease, patients should undergo regular tertiary care follow-up with functional assessment and advanced chest imaging.

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1. Introduction

Esophageal atresia (EA) with or without tracheoesophageal fistula (TEF) occurs in one per 3000 live births.^{1–3} In recent decades, improved surgical techniques and better neonatal supportive care have resulted in reduced mortality and increased prevalence of long-term disease-related complications, the most frequent of which include respiratory manifestations.⁴

Recurrent-to-chronic respiratory symptoms may upset daily life of EA survivors,⁵ and this is the reason why the assessment of pulmonary disease is recommended in these patients. This integrative review describes the various techniques currently used in pediatric clinical practice for assessing EA-related respiratory disease. Moreover, we present a novel algorithm for the evaluation and surveillance of lung disease in EA survivors. We carried out an electronic keyword literature search for English articles published on this topic up to September 22, 2015, in the Scopus, Web of Science, PubMed, and MEDLINE databases. We excluded the studies conducted exclusively on adults, but included those with a study population comprising children (or adolescents) and adults. The terms “esophageal atresia” AND (lung OR respiratory OR pulmonary OR airway or spirometry or complications or diagnostic tools) were used as keywords in combination, and the studies found were evaluated for selecting relevant literature. In addition, a manual search was conducted to evaluate review articles’ references. Literature reviews on diagnostic procedures for EA-related pulmonary disease prompted us to develop a novel algorithm for the evaluation and surveillance of lung disease in EA survivors.

2. Etiology

The etiology of pulmonary manifestations following EA repair is multifaceted. Because newborns with EA have an increased risk of premature birth that may initiate the clinical scenario,⁶ the association with anomalies such as tracheomalacia and lung hypoplasia may significantly contribute to respiratory morbidity since birth.² Gastrointestinal symptoms (i.e., regurgitation and/or feeding difficulties with repetitive cough during swallowing, and/or dysphagia and heartburn likely due to peptic esophagitis and Barrett’s esophagus) represent the major complaints at any age, and an association of gastrointestinal and respiratory symptoms has been hypothesized to imply a correlation between esophageal and lung dysfunction.^{1,5,7} Indeed, esophageal dysmotility and gastroesophageal reflux (GER) may cause and/or worsen wheezing, bronchial asthma, and pneumonia.⁸ Although the underlying mechanisms are still being debated, literature suggests that chronic asthma is likely elicited by a reflex mechanism and that recurrent pneumonia may be explained by repetitive acid aspiration.^{9,10} Chronic airway inflammation with bouts of infection can eventually result in segmental or even lobar damage leading to the development of severe, life-threatening lung disease in a proportion of patients.^{11–14} Finally, recurrent TEF may further complicate the clinical course.¹⁵ Following EA-TEF repair, structural anomalies persist in both the trachea and the esophagus, and chest

wall deformities, exacerbated by thoracotomy, may further contribute to alter pulmonary function.²

3. Respiratory complications

Patients with EA with or without TEF experience respiratory complaints more often and more persistently than other individuals, and recurrent bronchitis, chronic cough, repeated pneumonia, and asthma-like wheezing represent the major clinical manifestations.^{11,16}

Lower respiratory tract infection is abnormally common especially in the first years of life, with more than five annual respiratory tract infections and a rate of more than three attacks of bronchitis per year of up to 78%.^{17,18} In a study from Finland, aspiration pneumonia likely related to impaired esophageal peristalsis and esophageal stricture was reported in approximately 50% of affected children, although they did not experience more current respiratory or esophageal symptoms than those without.¹²

Coughs with sputum production and recurrent bronchitis are significantly more common among patients with repaired EA than among healthy individuals,¹⁹ and although respiratory morbidity tends to improve with age, chronic cough, associated with bronchial constriction and hyperresponsiveness, can persist or even become more frequent in adulthood.²⁰ As a consequence of repeated bouts of lower airways infection, bronchiectasis may also develop.²¹

Although some respiratory complications may be accounted for by documented tracheomalacia, esophageal dysmotility, GER disease (GERD), or surgical complications, a high proportion of EA survivors have abnormal pulmonary function that is apparently unrelated to these conditions.² A restrictive pattern generally prevailing over obstructive or restrictive-obstructive airway disease has been described in up to 96% of children, adolescents, and adults previously treated for EA with or without TEF.^{1,12,22–24} Interestingly, approximately one-third of a Finnish pediatric population had restrictive or obstructive defects that were apparently unrelated to current respiratory or esophageal symptoms.¹² In the same study, bronchial hyperresponsiveness was found to be severe/moderate or mild in 26% or 52% of the cases, respectively. Airflow obstruction may be explained by several mechanisms including small airway disease or proximal obstruction due to airway malacia or epithelial damage caused by GERD and recurrent episodes of bronchitis or aspiration pneumonia worsened by poor tracheal clearance, or decreased lung growth during infancy.²⁵ Multiple potential predisposing factors to restrictive lung disease are also congenital or acquired vertebral or chest wall abnormalities (i.e., scoliosis or postoperative rib fusions), surgical trauma, aspiration, and/or recurrent chest infections.¹

Chronic asthma is considered to be common in EA survivors, with significant bronchial inflammation also occurring in patients with nonallergic asthma.^{13,26} Whatever the initial trigger is, asthma significantly contributes to respiratory morbidity in EA, and it might even worsen pre-existing GERD.¹³

Table 1 Main findings from the 26 original articles that exclusively investigated respiratory disease in esophageal atresia survivors.

Study	Age (y)	Lung imaging	Pulmonary function tests	Bronchoscopy	Other	Main findings
Dudley & Phelan ¹⁸	1 to >9	—	—	—	Clinical outcome	Recurrent bronchitis during the first 3 y (78%)
Milligan & Levison ²²	7–18	—	Spirometry MCT	—	—	Obstructive (54%) & restrictive (21%) lung diseases
Couriel et al ³¹	8–17	—	Spirometry MCT	—	Clinical outcome	Bronchial hyperreactivity (65%) Bronchitis for > 8 y (25%) Mild restrictive lung disease
LeSouëf et al ²⁵	12–21	—	Spirometry Body plethysmography	—	—	Bronchial hyperreactivity (22%) Significant reduction of lung volumes in the pneumonia group vs. the nonpneumonia group
Chetcuti et al ³²	0–25	—	Body plethysmography	—	—	Daily cough (15%), wheezing (40%), bronchitis (34%) RV increase (77%), significant reduction in VC & FEV ₁ in patients who wheezed in the past 12 mo
Griscom & Martin ⁴⁰	2–21	CT	Spirometry	—	—	Bronchiectasis (40%) Mild restrictive (20%) & obstructive lung disease (40%)
Chetcuti et al ³³	6–37	—	Spirometry Body plethysmography	—	—	Reduced FEV ₁ (25%) & RV/TLC ratio (41%) Restrictive lung disease (18%)
Chetcuti & Phelan ¹¹	1–37	—	—	—	Clinical outcome	In the 0–5-y age group, pneumonia (50%), recurrent pneumonia (25%) Persistent cough (32% aged 0–5 y, 13% 5–10 y, 15% 10–15 y, & 9% > 15 y) Typical harsh cough (71% aged 0–5 y, 60% 5–10 y, 57% 10–15 y, & 40% > 15 y)
Beardsmore et al ³⁸	2–13 wk	—	Body plethysmography	—	—	Thoracic gas volume increase (33%), abnormalities in airway resistance pattern (78%), airway resistance increase (33%), limitation of inspiratory & expiratory airflow (11%)
Montgomery et al ³⁴	8–21	—	Spirometry Body	—	Clinical outcome	Asthma or bronchitis (39%)

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Table 1 (continued)

Study	Age (y)	Lung imaging	Pulmonary function tests	Bronchoscopy	Other	Main findings
			plethysmography Bicycle ergometer			Obstructive (44%) & restrictive (55%) lung disease Decreased maximal working capacity (53%)
Robertson et al ³⁵	7–28	X-ray	Spirometry Body plethysmography MCT	—	Clinical outcome	Respiratory symptoms (72%), bronchiectasis (4%), obstructive (12%), restrictive (36%), & mixed (4%) lung disease Positive MCT (24%)
Somppi et al ³⁶	3.5–30	—	Spirometry	Yes	Clinical outcome	Reduced FEV ₁ (67%) Tracheal inflammation (37%). Respiratory infections (29%), recurrent dyspnea (28%), & cough during the night (37%)
Agrawal et al ²⁶	7–12	—	Spirometry Body plethysmography	—	—	Restrictive lung disease (67%)
Choudhury et al ¹⁴	0 d to >30 d	—	—	—	Clinical outcome	Aspiration & pneumonia (13%) as early death causes Aspiration, tracheomalacia, & reactive airway disease as late death causes (59%)
Soto et al ³⁰	1–15	—	Spirometry	—	—	Restrictive lung disease (50%)
Little et al ²⁸	18.7	—	—	—	Clinical outcome	Respiratory infections (29%)
Sarnelli et al ¹³	0.8–14.6	X-ray HRCT Perfusion scintigraphy	—	Yes	—	Family history of atopy (40%) & allergic asthma (10%), lobar consolidations (80%), bronchiectasis (20%)
Banjar ²¹	1.25 ± 2.4	CT	Spirometry Body plethysmography	—	—	Tracheomalacia (29%), bronchiectasis (17%), obstructive (7%), restrictive (20%), & mixed lung disease (7%)
Lilja & Wester ²⁹	1–20	—	—	—	Clinical outcome	At 16–20 y, frequent cough between (36%), impaired exercise capacity (20%), respiratory infections (40%), & shortness of breath (53%)
Malmström et al ¹²	9.7–19.4	—	Spirometry Histamine challenge test	Yes	FeNO, clinical outcome	Obstructive (30%) & restrictive (35%) lung diseases Bronchial hyperreactivity (78%) Current respiratory symptoms (44%),

Table 1 (continued)

Study	Age (y)	Lung imaging	Pulmonary function tests	Bronchoscopy	Other	Main findings
Gischler et al ¹⁷	5–6.5	—	Spirometry Treadmill test	—	FeNO, clinical outcome	wheezing (52%), pneumonia (52%) Mild (72%) & moderate bronchitis in biopsies (7%) Abnormal FeNO (23%) Reduced FEV ₁ (25%) Abnormally low maximal exercise tolerance (6.3%) Normal FeNO High proportion of patients with >5 respiratory tract infections in 5 y (74%) Reduced Rrs6 (27%) & Rrs8 (24%) FEV ₁ significantly lower in EA with TEF vs. healthy controls
Harrison et al ³⁷	7.6 ± 2.2	—	Spirometry Forced oscillation technique Body plethysmography	—	—	Obstructive lung disease (13%) FVC & TLC significantly lower in EA with TEF vs. patients with gastroesophageal reflux disease
Peetsold et al ⁸	13.2 ± 2.9	—	Spirometry Body plethysmography Cardiopulmonary exercise testing	Yes	—	Obstructive lung disease (19%) Chronic cough (19%) & dyspnea (37%) Obstructive (50%) or restrictive (11%) lung disease
Spoel et al ³⁹	24–66 wk	—	Body plethysmography	—	—	Bilateral opacities, right lower lobe infiltrate, or pleural abnormalities (10%) Obstructive (19%) & restrictive (23%) lung disease Bronchial hyperreactivity (39%) Reduced ventilation reserve (45%)
Legrand et al ²³	13.3	—	Spirometry	—	Clinical outcome	
Beucher et al ²⁴	8.5	X-ray	Spirometry MCT Bicycle ergometer	—	—	

CT = computed tomography; EA = esophageal atresia; FeNO = fractional concentration of exhaled nitric oxide; FEV₁ = forced expiratory volume in one second; FVC = forced vital capacity; HRCT = high-resolution computed tomography; MCT = methacholine challenge test; Rrs6 = resistance at 6 Hz; Rrs8 = resistance at 8 Hz; RV = residual volume; TEF = tracheoesophageal fistula; TLC = total lung capacity; VC = vital capacity.

4. Management

With improved patient survival due to better neonatal care and surgery, the importance of recognition and management of pulmonary disease has increased. Table 1 summarizes the main findings from 26 original articles that exclusively investigated respiratory disease in EA survivors.

Several studies focused only on clinical outcomes,^{27–29} whereas others also included functional assessment by spirometry,^{30–38} airway challenge tests,^{12,24} and/or lung volumes measurement.^{8,39} The restrictive pattern was generally reported to prevail over the obstructive or mixed ones, and, overall, bronchial hyperresponsiveness was found in up to 78% of patients.¹² Of note, there were a few

studies on chest imaging findings, which were reported in only five articles.^{13,21,24,35,40} In particular, three chest computed tomography (CT) studies detected bronchiectasis^{13,40} and irregular cross-sectional shape of the trachea²¹ in a subgroup of patients, respectively. Chest CT findings may also include consolidations and/or bronchiectasis (Figure 1). By contrast, diffuse bronchial thickening, consolidations, and pleural abnormalities were the main chest X-ray findings described in a minority of patients.^{13,24,35} Airway endoscopy was seldom included in the available studies, with tracheomalacia representing a common finding^{8,12,41} and tracheobronchial inflammation being described in a variable proportion of cases.^{12,36} In addition to tracheomalacia and bronchomalacia, less common anatomic abnormalities may include ectopic or absence of bronchus and congenital bronchial stenosis.¹ Undoubtedly, evaluation of the airways structure via flexible bronchoscopy can help identify these problems in infants and children before EA/TEF repair, or also in those with persistent respiratory symptoms after EA/TEF surgery.^{12,13}

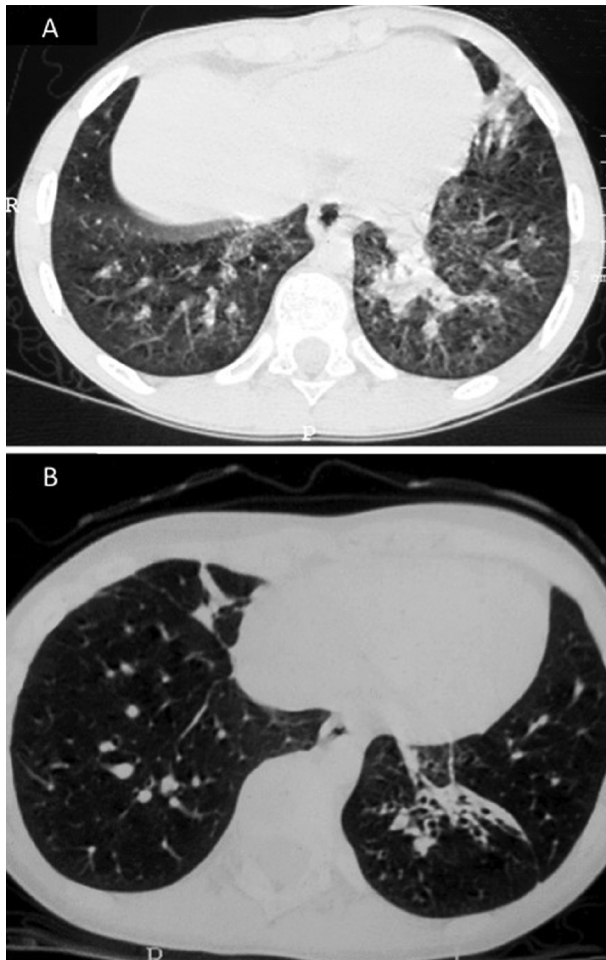


Figure 1 Chest high-resolution computed tomography from two children with esophageal atresia and tracheoesophageal fistula repaired at birth and followed at our department: (A) left lower lobe consolidations and (B) left lower lobe consolidation with bronchiectasis.

Although several articles on the main long-term respiratory complications in EA repair survivors have been published, a shared executive protocol has never been developed, nor have the available diagnostic tools been ordered in a management algorithm. Indeed, the few studies regarding the practical management of pulmonary complications in EA survivors and their heterogeneity make the development of an evidence-based operative algorithm virtually impossible. Nevertheless, due to the severe chronic complaints that some patients may experience and the impact on the healthcare costs, we propose a novel synthetic management algorithm (Figure 2), which may be helpful for clinicians dealing with lung disease secondary to EA. Basically, procedures are selected on the basis of the current clinical features. Present literature does not specify the timing of both follow-up visits and functional/chest imaging work-up. We suggest that basic procedures including transcutaneous pulse oximetry (SpO₂), chest radiographs, and lung function tests (the latter only on cooperating patients) are at least obtained in all patients at baseline. We also propose that EA survivors, with or without TEF, should be differentiated between those with mild airway disease and those with moderate-to-severe airway disease. In particular, patients with respiratory symptoms (persistent cough, recurrent-to-persistent wheezing, recurrent respiratory infections) who show slight abnormalities or normal results of SpO₂ at rest (ranging from 90% to 93%),^{42,43} and/or chest radiography, and/or spirometry (i.e., forced expiratory volume in 1 second and forced vital capacity $\geq 70\%$ predicted)⁴⁴ are defined as having mild airway disease. Conversely, patients with respiratory symptoms and more relevant abnormalities of SpO₂ at rest, and/or chest radiography, and/or spirometry are defined as having moderate-to-severe airway disease. We suggest that only the latter cases undergo regular tertiary care follow-up, including more extensive lung function assessment and advanced chest imaging (i.e., high-resolution CT and/or magnetic resonance imaging). A complete diagnostic approach to long-term respiratory complications after EA should also include tracheobronchial endoscopy with instillation of methylene blue for excluding recurrent TEF.⁵ Recurrent TEF should be corrected using laparoscopic antireflux procedures to prevent lung damage.^{45,46} These considerations lead to the conclusion that the evaluation of these patients is most efficiently accomplished in a tertiary care center where pediatric pulmonologists, gastroenterologists, radiologists, and surgeons are all available.

5. Conclusion

In patients following EA repair, recurrent-to-persistent respiratory disease represents a major feature, especially in early to middle childhood.⁴⁷ The persistence or recurrence of troublesome clinical manifestations imposes a scheduled follow-up of a large proportion of EA survivors, ideally through a multidisciplinary care approach for addressing their special needs. Pulmonary care of these patients involves managing comorbidities and preventing or minimizing damage to the lungs. Early detection and management of aspiration and other causes of recurrent-to-persistent lower airways infections in this population may

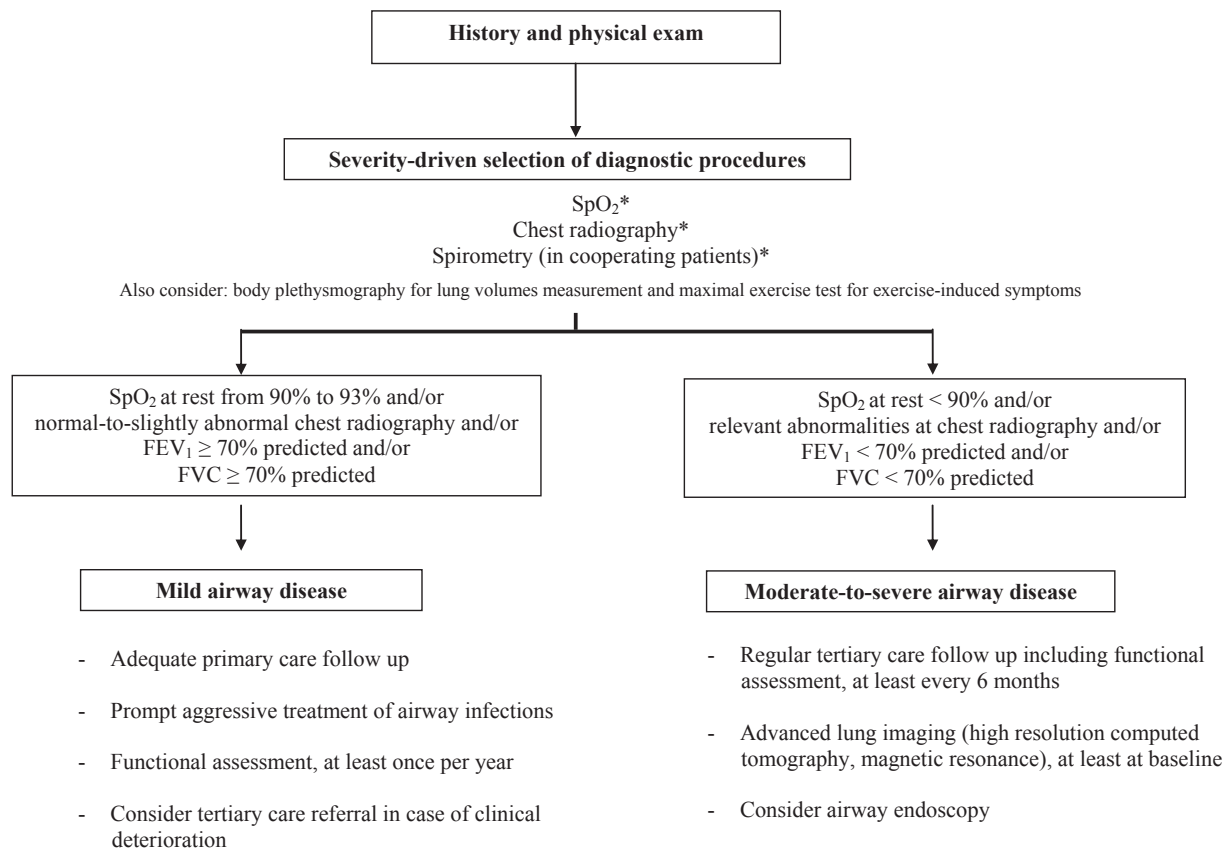


Figure 2 Algorithm for the evaluation and surveillance of chronic pulmonary manifestations in esophageal atresia survivors with or without tracheoesophageal fistula. * To be obtained in all patients. FEV₁ = forced expiratory volume in 1 second; FVC = forced vital capacity; SpO₂ = arterial oxygen saturation measured by pulse oximetry.

be important to prevent decrements in pulmonary function and serious long-term complications.³ A management algorithm for the evaluation and surveillance of EA-related respiratory disease based on the evidence from literature review is proposed. Like all algorithms, it is not meant to replace clinical judgment, but it should rather drive physicians to adopt a systematic approach to chronic pulmonary manifestations in EA survivors.

Ethical statement

This article does not contain any studies with human or animal subjects performed by any author(s).

Conflicts of interest

There are no financial or other relations that could lead to a conflicts of interest.

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